



# 'Articulations in Pediatric Rheumatology'

## The 1<sup>st</sup> National Conference in Pediatric Rheumatology 2003 Proceedings

Jointly hosted by:

**The Rheumatology Chapter of IAP,  
Mumbai Branch of IAP  
&  
Jaslok Hospital and Research Centre**



Venue  
Nehru Science Centre, Dr. E. Moses Road, Mumbai

Dates  
Sat. 8th November, 2003 & Sun. 9th November, 2003

# RCIAP 2003: The organizing team

Dr. Rachna Hasija  
Dr. T. Y. Kagalwala  
Dr. Raju Khubchandani

Dr. Roopa Srinivasan  
Dr. Pawan Sureka  
Dr. Naresh Thakkar

Dr. Vijay Viswanathan

Dr. Indu Khosla (MBIAP)

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Dr. Deepak Ugra (MBIAP)

## Acknowledgements

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*Compilation*

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**Dr. Raju Khubchandani**

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*RCIAP Logo*

**Dr. R. R. Navalkar,**

*Orthopedic Surgeon, Mumbai.*

## IAP Rheumatology Chapter

*Chair Person*

**Dr. Raju Khubchandani**

*Treasurer*

**Dr. Chetana Khemani**

*Secretary*

**Dr. Surjit Singh**

*Editor, Moves*

**Dr. Jyothi Raman**

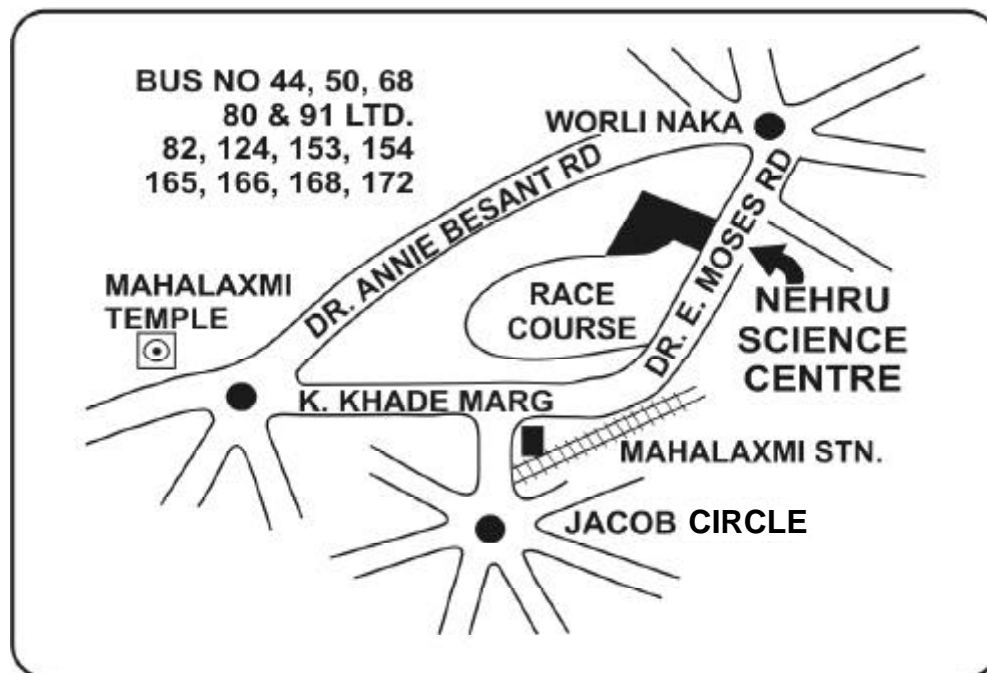
**Address:**

**Dr. R. P. Khubchandani, Kailash Darshan, 8th floor, Nana Chowk, Mumbai 400007.**

## IMPORTANT- NOTE CHANGE IN VENUE

The venue has been changed from  
Tata Hospital auditorium to  
**NEHRU SCIENCE CENTRE**

### HOW TO REACH NEHRU SCIENCE CENTRE



'This book has a limited print run. No copies will be available at the venue.  
You are thus requested to carry your mailed copy to the conference.'

## Greetings . . .



# Rheumatology Chapter - Indian Academy of Pediatrics

Ask any lay person about Pediatric Rheumatology and chances are that his bewildered look will say 'Arthritis? in children?' Ask a parent of a child with JIA or KD and his look will say 'We are lost' -we do not know where to go and what to do?' Their diametrically diverse responses summarize the status of this subspecialty – quantitatively small but qualitatively of immense significance.

In the recent years after the formation of the Pediatric Rheumatology Interest Group of the IAP there has been a visible interest shown by pediatricians in this niche area. This conference hopes to add fuel to this fire.

Internationally, Pediatric Rheumatology took birth in the mid 1940's. In the words of E. Bywaters, himself one of the pioneers in the field. " I think I can say that I saw it arrive, although I cannot specify its birthday or place and I am damned if I can read the father's signature on the birth certificate." It is hoped that this meeting at least marks its naming ceremony in the world's most populous country.

**R P Khubchandani**

*President*

RCIAP



## Jaslok Hospital & Research Centre

Jaslok Hospital & Research Centre (JHRC) a tertiary care multi speciality, charitable hospital was endowed to the city of Mumbai by Seth Lokoomal Chanrai, a philanthropist of high stature and foresight. It was inaugurated by the then Prime Minister of India, Smt. Indira Gandhi on 6<sup>th</sup> July, 1973.

Seth Chanrai envisaged and established an ultra-modern, 376 bedded, advanced centrally located medical institution with high level of expertise and technology of international standards. Its committed medical and paramedical staff have always strived to achieve their founder's dream. It has provided a model for similar other hospitals to be build in other cities of the country.

JHRC has over 25 specialities and sub-specialities and a full compliment of specialists of repute in various disciplines on its staff.

In keeping with the goal of striving for excellence and supporting medical events such as this one Jaslok Hospital is proud to co-host this event and wishes all the delegates a great learning experience.

**Col M Masand**

*Director General (Administration)*

Jaslok Hospital & Research Centre

## With best wishes from...

National President,  
Indian Academy of Pediatrics

It gives me immense pleasure to note that one of our youngest chapters has made rapid strides and is now holding a national conference with a distinguished international faculty. This sub-specialty has not received the attention that it deserved till now but fortunately the situation is changing.

The National Conference provides a unique opportunity to the participants to continue to learn, seek new information and knowledge and acquire and hone skills. Constant updating of knowledge and skills is imperative to be a good clinician and to provide quality care to our patients for the betterment of their health.

The program has been well chalked out to ensure that several important aspects are to be discussed in a very practical and unique format. I am sure the Conference will provide an opportunity to the delegates and faculty to interact and deliberate on several key issues.

Undoubtedly the Conference will fulfill the objectives with which it was conceived and I wish the event all the success.

Prof. H. P. S. Sachdev.

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President Elect  
Indian Academy of Pediatrics

I am glad to know that IAP Rheumatology group is organising the 1st National Conference is bringing out a compilation of interesting case studies for wider circulation. Pediatric Rheumatology has never received the importance that it deserved and hence I congratulate Dr. Khubchandani for taking the new initiative.

Dr. M. K. C Nair

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Hony. Secretary General  
Indian Academy of Pediatrics

It gives me pleasure to note that First National Conference of IAP Rheumatology Chapter will be held at Mumbai on 8th & 9th November 2003. The IAP Rheumatology Chapter, though a new entrant in IAP as well as in the pediatric

## With best wishes from...

specialties, is very active right from its inception. It is also an exciting pediatric specialty about which our members know very little and they will be keenly interested in learning more in this field. CMEs and Conferences are very important to learn a lot in a short time. It also gives opportunity to the delegates to interact with the experts. I am impressed by the format of the conference which will have more stress on discussion based on case scenarios and less of didactic lectures. The compilation of the proceedings will also help the delegates to refer to when in difficulties. I wish you all the best for the conference.

Dr. Nitin. K. Shah

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President

APLAR (Asia Pacific League Of Associations For Rheumatology)

May I have the privilege to send this message of goodwill and good wishes to our colleagues in the realm of Paediatric Rheumatology. We are delighted that you are holding your first national conference with an international flavour.

Paediatric Rheumatology is in infancy in India although a few rheumatologists have been trained in the discipline in other countries. I am happy to announce that a formal training programme in Paediatric Rheumatology has now been launched by Dr. Prudence Manners in Perth, Australia with positive support from APLAR.

In all our conferences Paediatric Rheumatology receives considerable attention and support. It is wonderful to see this discipline sprout deep roots, thanks to the tremendous efforts of Dr. Raju Khubchandani, Dr. Surjit Singh and Dr. Sujata Sawhney. This is just the beginning. More is certain to follow in a positive direction.

On behalf of APLAR, I wish the forthcoming national conference great strides.

Dr. P. K. Pispati

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“ A child with a rheumatic disorder needs specialised care. Are we well equipped? It is such endeavours that will provide us with the necessary skills.”

Dr. V. R. Joshi  
Senior Rheumatologist  
Mumbai

## PROFILING OUR INTERNATIONAL FACULTY



### **Dr. Angelo Ravelli, MD**

Age : 47 years Degrees -1985: Specialised in Pediatrics; 1988: Specialised in Allergology

Institution : IRCCS Istituto G. Gaslini, Pediatria II, Genoa, Italy

Achievements : 106 international articles

- Associate Editor of the Pediatric Rheumatology Online Journal (PROJ) and Clinical and Experimental Rheumatology.

Interests: Socio-cultural: politics, history; Sports: soccer, skiing" I am looking forward to visiting India and meeting a committed bunch of pediatricians who share a common love with me."

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### **Dr. Kevin Joseph Murray**

Age : 41 years

Affiliation : Senior lecturer, Dept. of pediatrics, School of Medicine and Dentistry, University of Western Australia.

Publications : 33

Presentations: 14 (international arena)

Your proudest moment in Pediatric Rheumatology:

Publication of 1<sup>st</sup> peer- reviewed paper

Interests : Sports: rock climbing, cricket, running and choir singing

"Rheumatology has for too long been an orphan speciality in pediatrics, and this conference is an opportunity to highlight the genuine place of rheumatology amongst the sub-specialities in pediatrics. In this International Bone and Joint Decade, we recognise the enormous impact of rheumatic and musculoskeletal disorders in adults, most of which have their genesis in childhood. I am delighted to be able to join with you in sharing some of my practical experience and ideas about these disorders."



## PROFILING OUR INTERNATIONAL FACULTY



### **Dr. Rolando Cimaz**

Age : 41 years

Affiliation : University of Milano, Italy

Publications : 125 abstracts and meeting proceedings, 95 full length papers

Presentations: 20 (half in the international arena)

Your proudest moment in Pediatric Rheumatology:

Fellowship in Dallas (1993), visiting Barbara Ansell, prize- best abstract ACR Boston-'98

Interests : volley ball (previous professional player- 1<sup>st</sup> Italian league), travel (especially to India) and to play with his two children. " Hope you enjoy this meeting and get interested in this fascinating subspeciality"

### **Sue Michelle Maillard**

Age : 35 years

Affiliation : Great Ormond Street hospital for children, London, U.K Degrees -MSc, MCSP, SRP

Publications : 4

Presentations: hundreds!

Your proudest moment in Pediatric Rheumatology:

My masters degree in Research methods and Statistics and completing my research in JDM.

Interests : spending time with my family, caring for my pets, ball room and Latin American dancing, volunteering for "The Children's Chronic Arthritis Association". "I have many years of experience in the field of physiotherapy in Pediatric Rheumatology, I hope to offer a slightly different perspective in the assessment and management of musculoskeletal symptoms. My focus will be on muscle strength and regaining full physical function using non-medical intervention "



# CONFERENCE PROGRAMME

## SAT, NOVEMBER 8<sup>TH</sup>, 2003

### REGISTRATION & BREAKFAST

**8:00 AM- 9:00 AM**

Topic	Speaker/Discussant	Duration	Page No
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#### SESSION 1:

1. Joint Examination	Sue Maillard	9:15 AM-10:00 AM	
2. Overview of symptoms in Pediatric Rheumatology	Sujata Sawhney	10:00 AM-10:30 AM	
3. Approach to a child with musculoskeletal pain of a few days duration	Rolando Cimaz	10:30 AM-10:50 AM	
Illustrative cases with panel discussion	R. Khubchandani B U Pai	10:50 AM-11:30 AM	<b>10</b>

### COFFEE BREAK

**11:30 AM-12:00 PM**

4. Approach to a child with musculoskeletal pain of a few weeks duration	Kevin Murray	12:00 PM-12:20 AM	
Illustrative cases with panel discussion	Sujata Sawhney	12:20 PM-1:00 PM	<b>16</b>

### LUNCH BREAK

**1:00 PM- 2:00 PM**

#### SESSION 2:

1. Approach to a child with musculoskeletal pain of a few months duration	Kevin Murray	2:00 PM-2:20 PM	
Illustrative cases with panel discussion	R. Khubchandani B U Pai	2:20 PM-3:00 PM	<b>22</b>

### COFFEE BREAK

**3:00 PM- 3: 30 PM**

2. Musculoskeletal pain- treatment options	Kevin Murray	3:30 PM-4:30 PM	
3. Role of physiotherapy	Sue Maillard	4:30 PM-5:00 PM	

# CONFERENCE PROGRAMME

## SUN, NOVEMBER 9<sup>TH</sup>, 2003

### BREAKFAST

**8:30 AM**

Topic	Speaker/Discussant	Duration	Page No
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#### SESSION 3:

- |  |                                |                    |           |
|--|--------------------------------|--------------------|-----------|
| 1. Lupus - diagnosis and suspicion       | Angelo Ravelli                 | 9:00 AM- 9:20 AM   |           |
| Illustrative cases with panel discussion | Sujata Sawhney<br>Pawan Sureka | 9:20 AM- 10:00 AM  | <b>36</b> |
| 2. Management of lupus                   | Sujata Sawhney                 | 10:00 AM- 10:30 AM |           |
| Illustrative cases with panel discussion |                                |                    | <b>36</b> |
| 3. Osteoporosis                          | Rolando Cimaz                  | 10:30 AM- 11:00 AM |           |

### COFFEE BREAK

**11:00 AM- 11:30 AM**

- |   |                                |                    |           |
|---|--------------------------------|--------------------|-----------|
| 4. Vasculitides - spotting Kawasaki disease | Angelo Ravelli                 | 11:30 AM- 12:30 PM |           |
| Illustrative cases with panel discussion    | Surjit Singh<br>Naresh Thakker | 12:30 PM- 1:00 PM  | <b>46</b> |

### LUNCH BREAK

**1.00 PM- 2.00 PM**

#### SESSION 4:

- |   |              |                   |           |
|---|--------------|-------------------|-----------|
| 1. Dermatomyositis - a clinician overview | Kevin Murray | 2:00 PM - 2:20 PM |           |
| Illustrative cases with panel discussion  | Surjit Singh | 2:30 PM- 3:30 PM  | <b>66</b> |

### COFFEE BREAK

**3:30 PM- 4:00 PM**

- |  |                 |                  |           |
|--|-----------------|------------------|-----------|
| 2. Rheumatic fever                       | Rolando Cimaz   | 4:00 PM- 4:30 PM |           |
| 3. Pain amplification syndromes          | Kevin Murray    | 4:30 PM- 5:30 PM |           |
| Illustrative cases with panel discussion | R. Khubchandani |                  | <b>78</b> |

## Session : 1

### CASE 1

Baby **S**, aged fifteen months, was brought to the casualty with complaints of moderate to high-grade fever since 6 days, associated with increasing irritability.

Her mother had noticed that **S**. hardly moved her right lower limb and cried if it was touched. She had stopped weight bearing and wanted to be carried all the time.

There was no history of trauma nor was any swelling noticed. There were no respiratory or GI complaints.

#### Questions I:

1. What is your first impression?
2. Why there is special mention regarding respiratory or GI complaints?

Surprisingly, **S**'s mom too had frequent non-specific musculoskeletal pains on and off since her delivery. She had a history of a fracture of her left arm. A physician had performed an RF test six months ago and it had been reported as weakly positive. She had been on anti-inflammatory drugs intermittently and had also received some pain killer injections. Due to her complaints, she found it difficult to carry her baby.

Baby **S** was her first-born, full term normal delivery, and she had been exclusively breast fed for 7 months after which a weaning diet had been started.

#### Questions II:

1. How would you interpret the mother's 'weakly positive' RF test and would you associate it with **S**'s condition?
2. What do you think is the most likely problem with mom?

We found her well grown for her age. (Weight = 10 kgs)

She was very irritable and had a heart rate of 140/ minute.

General examination revealed that there was evidence of rickets. **S** resisted being touched in the region of her right lower limb. She kept her leg in an attitude of flexion at right knee and there was some degree of warmth in the area of the knee.

Systemic examination revealed a hepatomegaly of 3- 4 cms and a just palpable spleen.

Investigations revealed a leucocytosis, positive CRP and an ESR of 50 mm/hr.

#### Questions III:

1. What other investigations would you send? Would you tap the knee?
2. Would you consider any kind of imaging/scan and if so which modality?

In view of fever, high counts and positive CRP, IV antibiotics were started.

Within 48 hours of IV antibiotics, fever spikes reduced and her limb was less tender. She was attempting to support on day 3 Her blood culture grew a common pathogen, though uncommon for her age.



## Questions IV:

1. What antibiotics would you select at onset?
2. The blood culture grew an uncommon pathogen. What do you suppose it grew?
3. What condition may be a predisposing factor for this kind of illness?

(Case compiled by Jyothi Raman, **Jaslok Hospital, Mumbai.**)

## CASE 2

**DK**, 11 year-old female, experienced a sudden 'tearing' sound in her left knee, while playing kho-kho. In a couple of hours, she was found to have pain and swelling of the left knee joint. Her family doctor noticed some swelling of the left knee and she was treated with some warm fomentation and NSAIDs. She was better in a few days and no further action was taken. Since then however she complains of pain in the left knee towards the evening. There is also a history of 'sudden giving in' which leads her to buckle in while running or walking.

## Questions:

1. What could be the probable causes of her problem?
2. What should the family doctor have done?
3. What are you likely to look for or find on clinical examination?
4. Would you investigate her and if so how?

(Case compiled by **Taruna Vohra**, Jaslok Hospital, Mumbai.)

## CASE 3

8 year-old **A** presented with complaints of fever and red eyes since 5 days and restricted movement of left wrist joint since one day. A week prior to these symptoms she had loose motions with blood and mucus for 2-3 days that was treated with nalidixic acid. Findings on examination were fever, puffiness of eyes, swelling of the left wrist joint with decreased range of movements (ROM).

## Questions:

1. What is your differential diagnosis?
2. What tests will help confirm your diagnosis?
3. How would you treat this patient?
4. The father is known to be seeing a rheumatologist for a long-standing 'back problem' with arthritis of his right knee and ankle? Any connection?
5. What will you advise the parents /How will you counsel them regarding the future?

(Case compiled by **Taruna Vohra**, **Jaslok Hospital, Mumbai.**)



## CASE 4

**A** was born as a healthy 3.7 kg to non- consanguineous parents who hailed from Gujarat. He did reasonably for the first 2 years of his life and then had some episodes of vague 'body-aches' when he was 2½ years. At 3 years he developed severe pain and movement restriction of his right shoulder joint for which he was admitted to hospital. Investigations revealed a polymorphonuclear leucocytosis with an ESR of 48 mm at the end of an hour. The X-Ray was reported (?) to show widened joint space. He was treated with NSAIDs and antibiotics. No diagnosis was offered and he recovered completely in about a week.

## Questions I:

1. At this stage, what would be your thoughts?
2. What do you think he was treated as?

4 months later, there was fever and severe abdominal pain. At that time he was noted to have some pallor. Acute appendicitis was considered but again he responded to antibiotics and anti-spasmodics. Another 6 months later, he suffered from pain, tenderness and inflammation of the upper tibial region. Symptoms recurred 2 months later, as pain and tenderness on the dorsum of both feet. Investigations revealed a Hb of 10 gm%, WBC of 8,800 with normal differentials and an ESR of 54. Records of his weight and height at this stage showed him to be on the 25<sup>th</sup> centile.

## Questions II:

1. Will you want to change your differentials?
2. What further investigations and history will you now ask for?

He was on and off NSAID's and a year later he had to be admitted for mild fever and investigations revealed a Hb of 9 gms, WBC count of 9,600 with 60% polymorphs. His X-ray showed a pneumonia that was treated with antibiotics.

8 months later he complained of chest pain, right knee and ankle pain. On examination there was warmth and swelling around the right ankle and left elbow. The investigations done at that stage were: ANA and RF - negative, ASO, PS for MP- negative, bone scan- multiple cold spots in vertebrae and lower end of femur, slide test for sickling was negative.

## Questions III:

1. How are this child's complaints typified - Acute? Chronic? Recurrent? Persistent?
2. What diagnosis are you considering now?
3. What test clinched the diagnosis?
4. Retrospectively what was the bone-scan telling us? What will you do to complete the work up?
5. Any new drugs to manage this condition?
6. How will you counsel the family and follow up the patient?

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai.**)



## Session : 1

### CASE 1

**AAKS** parents are worried. They have brought their 11 month-old son from Dubai.

He was a healthy full term with a birth weight of 3.5 kgs. He had been fine for the 1<sup>st</sup> 9 months. At that stage he had one episode of fever with sore throat that had settled with symptomatic treatment.

One week later, his high-grade fever recurred, accompanied with excessive crying. Examination by his pediatrician revealed a swollen hot right knee joint.

#### Questions I:

1. What is your differential diagnosis?
2. What other examination findings would you look for?

Blood investigations revealed polymorphonuclear leucocytosis and an ESR of 130 mm/hr. Just an anti-inflammatory drug was prescribed while awaiting test results.

#### Questions II:

1. Would you have started him on an antibiotic even while awaiting reports?
2. Would you have done a joint aspiration on him? How would you interpret the joint fluid results?

After staying well for a period of 2 days, fever and pain recurred. This time, the pain was localised to the left hip joint.

He was diagnosed to have septic arthritis on the basis of the above test reports and underwent knee and hip joint exploration for drainage of inflammatory fluid (sterile culture) and was started on multiple antibiotics in Dubai. After a week of therapy with no significant improvement, he was brought to our hospital for further evaluation.

On examination we found a happy, alert, active child with a scar over the right knee but no evidence of synovitis. The left hip continued to be painful. He held his hip in mid-flexion and was unable to bear weight on the left leg.

MRI of the knee and hip joint showed widening of the left hip joint space and the synovium was thickened and lit up well on MRI. Enhancing soft tissue and thickened synovium in the right knee joint was seen.

#### Questions III:

1. Do you wish to change the diagnosis or are you in agreement with a diagnosis of partially treated pyogenic arthritis that has been proposed to the patient? Reasons? Would you consider Oligoarticular JIA in the DD?
2. How will you interpret his MRI as a clinician and will this make a change in your treatment modalities?
3. When and how would you follow up this child?

**AAKS** was discharged on medication - happy, showing good growth after 6 weeks.



## Questions IV:

1. What discharge advise or tests will you suggest?

(Case compiled by Roopa Srinivasan, **Jaslok hospital, Mumbai.**)

## CASE 2

**VS**, 9yr old girl was seen for pain and swelling of joints since three months. The pain started in the right knee followed by left knee → left ankle → left shoulder → left wrist → small joints of hand and right wrist in that order. Pain was more intense in the morning and improved as the day progressed. The child was on nimesulide round the clock. There was no history of any rash, mouth ulcers or hairfall. Examination revealed pallor, generalised lymphadenopathy, mild hepatosplenomegaly, puffy small joints, painful movements of left wrist and minimal effusion of right knee joint. On investigation, she was found to have low hemoglobin (7.2 gm%), Leucopenia (4100/cumm), high ESR (133), RF was reported positive (80 IU /ml).

## Questions:

1. What differentials are you entertaining?
2. She has been referred as a case of RF positive polyarticular JIA. Agree or not?
3. Would you do an ANA in her? What would it tell you?
4. What test do you think clinched the diagnosis?
5. Was nimesulide her friend or foe?

(Case compiled by Taruna Vohra, **Jaslok hospital, Mumbai.**)

## CASE 3

**AAA**, 10yr old boy presented with swelling of the knee joint following a fall. Child has had such repeated episodes in the past. On two occasions the elbow joint was involved.

He was found to have a swelling of the right knee with a ten-degree flexion deformity of the left knee and decreased ROM at the left elbow joint. His acute phase reactants are normal.

## Questions:

1. What other contributory history would you enquire for?
2. What are the diagnostic possibilities and what specific investigations would you want in line with your thoughts?
3. Outline your management plan?

(Case compiled by Taruna Vohra, **Jaslok hospital, Mumbai.**)

## CASE 4

**D**, 2 ½ months of age, was admitted with a history of fever, acute painful swelling of right elbow and restriction of movement of shoulder and hip. He hailed from the land of the Taj,



Agra, where he had been treated with multiple IV antibiotics and with dexamethasone added (reasons?) after a non-responsive week.

In spite of treatment, migratory polyarthritis with fever was the dominant mode of presentation.

The only past history was an attack of acute gastroenteritis about 7 weeks before this episode. He had been hospitalised, and investigations documented prerenal azotemia and anaemia (Hb- 9.0 gm%) for which he was treated with IV fluids, antibiotics and a blood transfusion.

Questions I: Any thoughts?

To take the story forward, he was eventually referred to Mumbai for arthrocentesis ! On admission, he weighed 4.5 kg was irritable, febrile, had a swelling of right elbow and pain on movement of his right hip and shoulder. A day after admission, the left knee appeared puffy as did some of the small joints of the fingers.

Fever was high grade, continuous with 2-3 spikes per day.

Questions II: Would you do the arthrocentesis?

Polymorphonuclear leucocytosis, with high ESR and platelet count of 11 lakhs. HIV negative, SGPT, BUN, Creatinine were all normal.

- Questions III:
1. There is a specific point in his history that gives a clue. What do you think it may be? Hence what further investigations would you ask for?
  2. What are your views on his platelet count? Could this be an atypical Kawasaki disease?
  3. What treatment options exist at this stage? What NSAID's can you use in this age group?

(Case compiled by Jyothi Raman, **Jaslok hospital, Mumbai.**)

OVER TO SESSION 2.....



## SESSION : 2

### CASE 1

Three-month old, female child, born of 2nd degree consanguineous marriage, was brought with complaints of excessive crying, paucity of movements of limbs and dark patches noted over ankles. They said that there had been very little weight gain since birth.

#### Questions I:

1. What else would you like to know?

The unfortunate couple had lost their first son at 9 months of age due to very similar problems.

We noted that the fingers were held in flexion and there were contractures of distal PIP joints. There were palpable, subcutaneous nodules, particularly near joints.

#### Questions II:

1. Is this rheumatoid arthritis in a 3 month old?
2. What other specific clinical features would you look for in this very irritable child?
3. The diagnosis is striking and can be made at first glance. Would you be able to confirm it by some objective test?
4. How would you prognosticate the condition and when will you ask them to come back?

(Case compiled by Jyothi Raman, **Jaslok Hospital, Mumbai.**)

### CASE 2

She was the 1<sup>st</sup> case of arthritis I saw as a junior resident doctor in July 1999. **TG**, an eleven year-old child lay in bed, her shrivelled and delicate body frame spoke volumes of the rampage created on her health by her disease. Her parents had given up hopes, but I saw a quiet determination in **TG**'s eyes.

**TG** had been admitted for prolonged pyrexia of 1-year duration, initially low grade to start with, then progressing to high grade. There were joint swellings involving multiple large and small joints, inability to walk, puffiness of the face, patchy alopecia and mouth ulcers since 3 months.

There were several hospitalisations for this fever with various diagnoses of enteric fever, malaria and tuberculosis entertained along the way.

In the early part of the illness i.e. in 1998, there was an evanescent rash and abdominal pain that accompanied the fever. The rash was labelled as a drug rash by the treating physician.

Later in the course of her illness, she had deteriorated acutely with respiratory distress and had to be ventilated. She dramatically improved in 2 days with antibiotics and methylprednisolone; a 2D-echo had shown a small pericardial effusion.



Even after discharge, **TG** continued to spike fever and there was loss of weight and appetite. 2 months later, she had an episode of convulsions with high-grade fever and jaundice, her CT scan and LP were normal.

Questions I:

1. What more would you like to know about the fever?
2. Why did she suffer from acute respiratory distress and why did it dramatically improve?
3. What was the probable cause of the convulsion and jaundice?
4. Is the pericardial effusion significant? Was the drug rash truly so?
5. With this story what direction do you think you are headed?

On examination, **TG** had patchy alopecia, cervical and axillary lymphadenopathy, and hyperpigmentation of both her knees. She had generalised hypotonia and muscle wasting, there was decreased range of motion swelling and deformity of the large and small joints. On examination, we also found that there was firm hepatosplenomegaly and a soft systolic murmur.

Questions II:

1. Why was there patchy alopecia and hyperpigmentation around the joints?
2. Would lupus enter your differential diagnosis?

Investigations revealed the following:

Tests	16/4/98	20/4/98	8/5/98	20/7/99
Hb	8.5 gm%	9 gm%	8.5 gm%	5.96 gm%
TLC	14,500; P <sub>86%</sub>	45,600; P <sub>86%</sub>	28,600; P <sub>84%</sub>	27,600; P <sub>86%</sub>
Widal	Positive			
CRP				96
ESR				110
S.Ferritin				1745 (10-100ng/ml)
S. Ig				IgG & IgA ↑, IgM (n),
ANA, RF				Negative
XRC	Left pleural effusion	Bilateral pleural effusion	Patchy pneumonitis	

Questions III:

1. Low Hb with high ferritin. Explain?
2. Why was Widal positive?
3. Would you have treated her for tuberculosis since she has a pleural effusion?

She was started on treatment and monitored with relevant investigations and functional scales. **TG** gradually started walking and now goes to school. She is a bright-eyed girl with



grit and determination. Recently she developed a swelling near her left eye that has persisted. Her doctor is puzzled at this latest development.

- Questions IV:**
1. What was the therapy she could have been started on?
  2. What type of functional assessment scales are you aware of?
  3. What could be the cause of swelling near the eye?

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai.**)

### CASE 3

**DM** had come down to Mumbai from Oman. She was 4 years old but had the appearance of a 2 year-old with severe failure to thrive. When the consulting doctor saw her, she appeared withdrawn, sick and in obvious pain. She had a history of recurrent fever and swelling of joints for 2 years with multiple hospital admissions. During one such admission in 1999, a maculopapular rash, hepatosplenomegaly, cervical and axillary lymphadenopathy were noted. Parents had noted a micrognathia between 3 and 4 years of age.

- Questions I:**
1. What is your first diagnosis?
  2. While following up these patients, what specific tests or examinations would you perform?

**DM** was already on the appropriate treatment measures on admission, but her fever, pain, lymphadenopathy and hepatosplenomegaly persisted. Her joint examination revealed micrognathia and restriction of TM joint movement, restriction of neck movements, swollen, tender and warm wrists, elbows, knees and ankles and puffy PIP's. Her investigations revealed the following:

Tests	7/12/99	30/12/99	1/4/00	7/8/00	9/11/00(Jaslok)
Hb	9.2 gm%	8.2 gm%	8 gm%	7.9 gm%	7.7 gm%
TLC	28,300P <sub>82%</sub>	8,100	12,800;P <sub>70%</sub>	31,600;P <sub>80%</sub>	44,900 P <sub>68%</sub>
Platelets	6,25,000	2,53,000	7,84,000	5,50,000	8,71,000
ANA; RF		Negative			Negative RF
CRP		455	172		
ESR	52	59	110	71	34
LFT; RFT	NORMAL				
Eye					Normal
2D-echo		Normal			Normal
X-Ray		XRC- Rt para-cardiac patch			Knee→no erosion, TM jt→bilaterally hypoplastic; Cervical spine→no dislocation.



## Questions II:

1. After reviewing the CBC, what picture emerges?
2. Comment on the trend and the utility of ESR and CRP in chronic arthritis in children?
3. What is your thought about the platelet count? Does it have prognostic significance?
4. What do you think was wrong with the cervical spine?
5. Why was there micrognathia and would you advise surgery for the same?

**DM** improved with therapy in the hospital, she was afebrile towards the latter part of her hospital stay, her mood improved significantly and she started taking part in self-care activities.

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai.**)

## CASE 4

**SP**, 4 yr old female child, was noticed to walk with a limp by her parents when she was two years old. She was diagnosed as having traumatic synovitis of the left knee joint by an orthopedic surgeon and treated with NSAID's for a few weeks after which she did well. 2yrs later, she came with swelling of the right knee joint which had persisted for 9 months. O/E she was found to have swelling, effusion and restricted movements of the right knee joint.

## Questions:

1. What is your clinical impression?
2. What would you expect her ESR to be? Any other blood tests? Will you tap her joint? Will any sign make you change your mind regarding tapping the joint?
3. Would you need to monitor any other parameters? If so for how long?
4. What is the treatment of choice?

(Case compiled by Taruna Vohra, **Jaslok Hospital, Mumbai.**)

## CASE 5

**AG** is a 7-year-old from Bhillai. He is a known case of Juvenile Idiopathic Arthritis- Systemic Onset, diagnosed at the age of 3 years and 8 months. He is currently on oral Ibuprofen and Methotrexate. However, he gets a lot of vomiting and nausea due to the medications.

## Questions I:

1. How can you help him at this stage?
2. How would you monitor methotrexate Therapy?

In Jan 2003, **AG** suddenly deteriorated and his pain in the joints became more severe. There



was a frequent rise in temperature, high grade that required admission. His tests for finding an infectious cause were negative. His ANA and RA continued to be negative. His dose of methotrexate was further increased. His investigations were as thus:

Tests	31/3/00	30/1/02	17/1/03	21/1/03	12/2/03
Hb	12.8 gm%	12.9 gm%	10.6 gm%	9.5 gm%	8.3 gm%
TLC	12,100 P <sub>45%</sub>	9,200 P <sub>45%</sub>	14,000 P <sub>80%</sub>	36,500; P <sub>90%</sub>	36,800P <sub>80%</sub>
ESR	14	40		110	112

#### Questions II:

1. What is Aditya's problem and when did it begin?
2. What will you do to treat the problem?
3. How will you ensure that the problem's recurrence is avoided?

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai.**)

## CASE 6

A 12 year-old girl presented with h/o rash, fever and fatigue since 10 days and a diffuse swelling of the right upper limb was hospitalised. O/E, a faint generalised maculo-papular rash, mild pallor and pitting oedema of the right arm were present. Mild neutrophilic leucocytosis and elevated ESR were present on investigation. Her clinical picture rapidly deteriorated with progressive involvement of renal, cardiac and coagulation systems. Blood counts and ESR also dipped. A procedure done clinched the diagnosis.

#### Questions:

1. What do you think is the complication that occurred?
2. What procedure would have clinched the diagnosis?

"The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by Sureshkumar EK (Pediatrician) and Ramesh Bhasi (Rheumatologist) from **MIMS, Calicut, Kerala** and thanks them for their valuable participation."

## CASE 7

**S**, 7 years now, is a diagnosed case of SOJIA on oral steroids, NSAID's and Methotrexate. Patient was showing some improvement, when...

Day 12 → Multiple episodes of focal convulsions with altered sensorium and fever.

Investigations: CSF and CT Brain- Normal, PS for MP- P.Falciparum +

Patient was treated with antimalarials and showed improvement.

Day 22 → Repeated episodes of multifocal convulsions with altered sensorium and fever.

On examination: pallor ++, petechiae +, Liver 3 cms, Spleen just palpable.

Day 26 → Tachypnoea, icterus, P/A: Hepatosplenomegaly increased

Investigation chart:



Investigation	Admission	Day 12	Day 22	Day 26	Day 31
Hb	12.4	11.7	7	8	6.5
WBC	17,600	35,800	19,900	18,500	1600
Platelets	2,30,000	4,45,000	12,000	1,27,000	18,000
ESR	90	70	120	50	15
LFT	normal			Bil-8, SGOT-108 PT- 38	Bil-8, SGOT-234 PT- 23
PS for MP	Negative	P.Falciparum	P. Vivax	Negative	Negative

## Questions:

1. Correlate the clinical picture and investigation findings?
2. What further investigations would you suggest?
3. Are there any lab correlates with prognosis of this condition?

“The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by the Pediatric Team (S.Joshi, S. Save, S. Malik, C. Modi, A. Aiyar, A. Kochar, M. Ahirra, S. Tank) of **BYL Nair Hospital, Mumbai** and thanks them for their valuable participation.”

## CASE 8

5-year old **N** suffers from a peculiar problem. She suffers from recurrent pains in her fingers, thighs and ankles since 2 months which worsen in the evenings. Two years ago, she dislocated her left elbow. She is left-handed and complains of pain after writing at school and her teacher has said that this is a clear case of writer's cramp. On examination you find a small effusion in the knee. Her GP thinks she has school avoidance behaviour

## Questions:

1. Are you happy with his diagnosis?
2. Have you heard of Beighton's scoring?
3. How would you investigate her?
4. How will you treat her?

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai**.)

## CASE 9

**V**, a 6 year-old male came to us with the history of being unwell since the age of 2 years. At 2 years→ diagnosed with primary complex, given AKT. At 3 years→ otitis media. After 2 months→ Pneumonia, respiratory distress and pre-renal azotemia. Thereafter → Recurrent LRTI's, ear infections, arthritis at different sites. Presently→ monoarticular (knee) synovitis. Similar knee involvement in the past.

## Questions :

1. What conclusion do you draw from the presenting complaints?

(Case compiled by Roopa Srinivasan, **Jaslok Hospital, Mumbai**.)



## CASE 10

A 17-year-old girl, known case of Turner's syndrome, presented with complaints of difficulty in walking due to inability to flex her knee joints since 1 year, difficulty in flexion of elbow joints and bony swellings noticed over her wrists since 1 year, pain over small joints of the hand and occasional low-grade fever since 1 month. O/E: She had multiple, symmetrical large and small joint effusions. There was evidence of tenosynovitis. Her investigations revealed Hb 8.1gm%, WBC 9110, N<sub>60%</sub> L<sub>30%</sub>, ESR 120, Platelets 5,35,000, RF- POSITIVE (1:512)

## Questions:

1. Is the history of Turner's syndrome relevant?
2. What is your diagnosis?
3. What other clinical features will you look for in this child (with respect to the arthritis)?

(Case compiled by Jyothi Raman, **Jaslok Hospital, Mumbai.**)

## CASE 11

A 12-year girl from Madhya Pradesh presented in June 2002, with a history of joint pains and stunted growth. The parents gave a history of fever, off and on, since the age of 2-3 years. The arthritis involved the wrists, knees and ankles in the last 10 months. There was decreased vision since the last 9-10 months. Further questioning revealed – dryness of the mouth, recurrent skin rashes over the lower limbs and shortness of breath on exertion. In the past, she had been investigated for fever and given a course of anti-TB drugs.

## Questions I:

1. What are the thoughts running in your mind?

On examination, the child was febrile and pale. Pulses were normal. She stood 122 cm tall and weighed 19 kg. Red, nodular rash was noticed over the shins. There was swelling and minimal tenderness of both wrists, ankles. Bilateral knee effusions were present. There no muscle tenderness or weakness. Splenomegaly was present. CVS and RS examinations were normal. The CBC revealed- Hb 9.5gm%, WBC 3800/mm<sup>3</sup>, Platelets 3.6 lakh. ESR was 55 mm. Other investigations were: ANA 1+, Rheumatoid factor – negative, Mantoux test was negative. HLA-B27 was negative. Ophthalmological examination showed bilateral chronic iridocyclitis with band keratopathy.

## Questions II:

1. Name two investigations that would suggest the diagnosis?
2. How would you confirm the diagnosis?
3. How will you manage her?

“The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by Mala Bhambhani from **Mumbai** and thanks her for her valuable participation.”



## SESSION : 3

### CASE 1

A ten year old girl,

- Well till 2001 summer, developed symmetrical polyarthritis involving small joints of hands, wrists, elbows and knees.
- ESR 65, platelets 4.4lakhs/mm<sup>3</sup>, Hb 10.4gm%. RF negative.
- Diagnosed: Seronegative poly JIA - treated with methotrexate.

Presented to us: January 2002-low grade fever, lethargy, breathless on exertion.

- Examination:
  - Cachectic child
  - Weight 21 kg,
  - Febrile
  - Multiple small lymph nodes,
  - Polyarthritis
  - Hepatosplenomegaly

Investigated:

- ESR 100, platelet count 2.2lakhs, WBC 5600, CK 600 I.U./L, CRP negative.
- ANA: positive 1:160,
- Urine: 24-hour protein- 300mg.

#### Questions:

1. What are the clinical possibilities in this patient?
2. How would you interpret her investigations done so far?
3. What further investigations would you order at this stage?

### CASE 2

10 year old girl,

- Unwell since February this year.
  - Low grade fever
  - Aching limbs
  - Lost 3 kg weight.
  - Facial rash for the last one month.
- Poor socio-economic status. Brother treated for tuberculosis.
- Seen by primary physician - Mantoux-21 mm with vesicular lesions!
- Anti-tuberculous treatment: 20<sup>th</sup> October 2002
- Presented on 1<sup>st</sup> November with
  - Fever
  - Malar rash
  - Hepatitis – SGOT 2456 I.U/I
  - ESR 78, CRP negative.
  - Platelet count 48000/mm<sup>3</sup>
  - ANA 1:160
  - DsDNA- positive
  - C<sub>3</sub> decreased.
  - Urine- NAD.







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NOVARTIS

- Examination: confirms all of the above, non-deforming arthritis with livedo reticularis.
- Serology suggestive of lupus: ANA positive, anticardiolipin positive.

Questions:

1. Do you need any further clinical data?
2. What is the significance of the anticardiolipin positivity ?

## CASE 6

An infant -

2-month-old FTND, to a primi gravida mother.

- Well infant noted to have a facial rash at 6 weeks age. Clinical examination confirmed erythematous rash on the forehead and cheeks with systemic and developmental assessment unremarkable. CBC showed a platelet count of 90,000/mm<sup>3</sup>.
- Work up for sepsis, and congenital infections negative, No known maternal disease.

Questions:

1. What is the differential diagnosis in this case?
2. What further information do you want?
3. What is the long-term prognosis in this child?

“The 1<sup>st</sup> RCIAP National Conference acknowledges contribution of all the above cases 1 to 6 by ~~faculty member~~, **Sujata Sawhney**, from **Gangaram Hospital, New Delhi** and thanks her for her valuable participation.”

## CASE 7

**P**, a 12-year-old girl presented with

h/o a swelling in front of the neck of 3 months duration.

Examination- goiter, with no clinical signs of hypo/ hyperthyroidism.

Investigations- low T<sub>3</sub> & T<sub>4</sub> levels, TSH: 26U/L (N: 0.5-5)

A diagnosis of primary hypothyroidism was made and she was started on Eltroxin.

In the past 3 years, she has had recurrent episodes of facial puffiness and mild pedal edema.

One such episode (6 months back) that was investigated, revealed nephrotic range proteinuria, hypoalbuminemia and hypercholesterolemia with mildly elevated renal parameters. The renal biopsy was suggestive of Minimal change disease.

She responded to steroids and the renal functions normalised with conservative management.

Off steroids for 4 months, she came back with complaints of facial puffiness and pedal edema and nephrotic range proteinuria.



Questions I:

1. What is your clinical impression?
2. **Primary hypothyroidism** 1<sup>st</sup> diagnosed at 12 years of age' - comment?
3. What feature points away from the 'typical nephrotic' picture in her?
4. Which test will diagnose the cause for hypothyroidism in her?
5. How would you approach this case and complete the work-up?

The repeat renal biopsy was consistent with WHO Class II b lupus nephritis. Immunofluorescence revealed deposits of Ig M 3+ in the mesangium and periphery in a diffuse and granular pattern.

Questions II:

1. Comment on the discrepancy in both the biopsy reports?
2. Outline the management and prognosis of various stages of lupus nephropathy based on kidney biopsy?
3. What complications occur with lupus nephropathies?
4. Outline the pharmacotherapy and modalities of managing lupus nephropathy?

"The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by S. Karthik, **St. John's Medical College Hospital, Bangalore**, and thanks her for her valuable participation."

OVER TO NEXT PART OF SESSION 3...

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Franco India Advt (half page)



## SESSION : 3

### CASE 1

A 10 month-old infant presented with high-grade fever of 5 days duration. The fever started abruptly and ranged between 39- 41.5°C. There was no associated respiratory catarrh, cough or urinary complaints. He had become extremely irritable over the last couple of days and was refusing to accept feeds. He had developed diarrhoea the day before hospitalisation – the stools were watery, there being no associated blood or mucus. He had been seen by the family physician who diagnosed serous otitis media and prescribed amoxycillin and paracetamol.

The child had been completely well the week before. He had received his vaccinations as per schedule. The father was a business executive who was frequently away from home while the mother was a schoolteacher. He attended a crèche during daytime when the mother was away at work. He had an older sibling, a boy of 5 years, who had had streptococcal sore throat the week before.

#### Questions I:

1. What is your impression about this child's illness?
2. Should he be hospitalised?
3. What questions would you like to ask the mother?
4. What specific physical findings would you look for?

The child was hospitalised. He had a temperature of 40.5°C, the heart rate was 140/minute, respiratory rate was 40/minute and the blood pressure was 100/70 mmHg. He was extremely irritable and was resenting examination. The anterior fontanelle was bulging but pulsatile. There was a faint macular, erythematous rash more prominent on the trunk but also seen on the limbs. It was non-itchy. He had red, fissured and swollen lips. The eyes were congested and the cheeks were red. His hands and feet appeared swollen and somewhat tender to touch. Examination of the ears showed congested drums, with a blurred cone of light, air bubbles and a fluid level.

Liver and spleen were just palpable. Chest examination showed bilateral fine crepts and occasional rhonchi. Auscultation of the heart revealed a short systolic murmur in the left parasternal region. Examination of the central nervous system revealed an inconsolable, extremely irritable child. One observer had noted a doubtful seventh cranial nerve palsy at admission but this finding could not be confirmed later. The deep tendon jerks were normally elicitable and the plantars were downgoing.

#### Questions II:

1. What are your clinical possibilities now?
2. How would you investigate this child?

The following investigations were carried out:



Day of admission	Hb(gm%) WBC count	WBC(/mm <sup>3</sup> ) count	Differential blood film	Platelet	ESR	Peripheral
0	11.8	22,000	P <sub>85</sub> L <sub>13</sub> E <sub>2</sub>	400,000	40	Normocytic, hypochromic
7	11	18,500	P <sub>76</sub> L <sub>22</sub> M <sub>1</sub> E <sub>1</sub>	620,000	84	Normocytic, normochromic
14	12.4	20,800	P <sub>80</sub> L <sub>18</sub> E <sub>2</sub>	1280,000	76	Normocytic, normochromic

Blood culture- Sterile; X-ray chest- Streaky infiltrates right parahilar region;

ASO < 100 U/ml (N = < 140); CRP = 4500 ng/ml (N = < 1500)

Throat swab culture – No pathogenic organism

Biochemistry

Day	Na+ Mmol/L	K+ Mmol/L	BUN mg%	S. Cr. mg%	Total proteins(A/G)	Bilirubin	AST < 55	ALT (45)	Alk PO4 (< 420)
0	135	4.5	30	0.8	6.2 gm% (4.2/2)	2.3 mg% (C=1.2)	69	58	280
5	130	4.8	28	0.7	6.0 gm%	1.8 mg% (C=1.4)	80	70	240

Urine examination

Day	Albumin	Glucose	Pus cells	RBCs	Casts	Culture
0	Traces	Nil	0-1/hpf	Occasional	Occasional	Sterile
5	1+	Nil	5-6/hpf	Nil	Nil	Sterile

**Ultrasound examination:** Abdomen – Liver showed normal echotexture; gall bladder appeared to be dilated and filled with thin fluid; spleen was enlarged; both kidneys were normal in size. Head – normal sized ventricles; mild subdural effusion.

**Question III:** How do you interpret these findings?

The residents looking after the child wanted to do a lumbar puncture for examination of cerebrospinal fluid but the attending consultant deferred the decision. He was scheduled to undergo a CT scan of head but this investigation was also deferred.

**Questions IV:** What would you have done in this situation?

A possibility of early meningitis was considered by the first on call and ceftriaxone was started.

**Questions V:** Do you agree with this line of management?



The case was discussed in the ward rounds. Some of the residents felt that there was evidence of a bacterial infection in view of the neutrophilic leukocytosis and the high-grade fever. Others felt that the child probably had a viral infection (? Infectious mononucleosis) and needed further observation. Fifth Disease was also considered in the differential diagnosis. A possibility of Kawasaki Disease was also considered at the time of admission and the attending consultant wanted to give a bolus of intravenous immunoglobulin (IVIG). The other consultant had a different opinion as he felt that the diagnosis of KD was still not certain and giving treatment at that stage was not warranted.

An echocardiographic examination was carried out at this time that was normal.

**Question VI:** What are your comments?

The debate continued and it was decided to defer IVIG therapy for the time being. Intravenous ceftriaxone was started. The fever persisted and the child continued to have irritability. By this time the child had had fever for almost 8 days. Repeat blood and urine cultures were sent and combination therapy with cefpodoxime/ amikacin was started. Blood culture revealed a growth of *Staphylococcus epidermidis*. Even though this was thought to be non-pathogenic, a trial of vancomycin was given. The fever, however, persisted.

**Question VII:** Is this a common clinical scenario in pediatrics?

By the 12<sup>th</sup> day when the fever had still not subsided, it was decided to give a bolus of IVIG empirically. A dose of 2 gm/kg was given in the morning. By the evening the fever had almost disappeared and the irritability had markedly decreased. The next day it was noticed that he had a peculiar peeling of the skin of fingers and toes. The parents were greatly alarmed at this phenomenon. A final diagnosis of Kawasaki Disease was made and the child was discharged on low dose aspirin.

- Question VIII:**
1. Was the unit justified in delaying therapy with IVIG in this child?
  2. What are the consequences of delayed therapy?
  3. What should one do if one is not certain of the diagnosis?

It is often said that thrombocytosis is a rather typical finding in children with Kawasaki Disease.

- Question IX:**
1. Do you see thrombocytosis in all patients with KD?
  2. In a clinical setting of KD, can one use the presence of thrombocytosis as a confirmatory test for KD?

Similarly, a lot of importance has been given to the occurrence of desquamation in KD.

**Question X:** Is peripheral desquamation virtually pathognomonic of KD?

The parents of this child were very worried regarding the long-term complications of KD.



**Question XI:** How would you counsel the family?

There is some evidence that KD may have cardiac sequelae much later in life. In fact childhood occurrence of KD has been mentioned as one of the causes of sudden death in adults.

**Question XII:** What are your comments?

According to one of the standard textbooks KD is the commonest vasculitic disorder in children. It is believed to be even commoner than Henoch Schonlein Purpura – a condition that is seen ever so often in children.

**Question XIII:** What is your opinion?

It is our impression that KD is being grossly under-diagnosed in children in India. Very few centers are regularly reporting this condition.

**Question XIV:** What is the situation in worldwide?

The older sibling of this child had developed a streptococcal sore throat prior to the onset of fever in the index patient.

**Question XV:** Do you think this had a bearing on the development of KD in this child?

KD is diagnosed on the basis of a set of clinical criteria that were first enunciated in the late eighties. There is, however, still lot of debate regarding the validity of these criteria. For instance, the Japanese workers have their own criteria.

**Question XVI:** What difficulties do you encounter in your day-to-day practice because of such ambiguities?

The dose of IVIG that has been used in KD has varied from 200 mg/kg/day for 4 days as in Japan to as much as 2 gm/kg single pulse as in the United States.

**Question XVII:** What are your views on this aspect of treatment of KD?**CASE 2**

**P**, a 6 year-old boy suddenly took ill. He presented with severe colicky pain in the abdomen for the last 12 hours associated with passage of black tarry stools. There was history of fever 3 days ago that had subsided on its own. The child was seen by his family physician who noted some abdominal distention, sluggish bowel sounds and tenderness in the right iliac fossa. He kept a clinical possibility of Meckel's diverticulum / acute appendicitis and referred



the child to hospital for surgery. The child was seen by the pediatric surgeons and a diagnosis of "Acute Abdomen" was made. Urgent ultrasound examination was done – it showed an ileo-colic intussusception along with some swelling of the appendix. A barium-assisted reduction was attempted but this was unsuccessful. Meanwhile the symptoms worsened and the child was kept for an emergency laparotomy.

**Question I:** What are your clinical possibilities?

During the pre-anaesthetic examination, a faint macular rash was noted over the feet and legs that were urticarial at places. A pediatric medicine consultation was sought. The resident kept a clinical possibility of a drug rash (?amoxicillin induced) and prescribed antihistaminics. Surgery was deferred for the time being.

**Question II:** What are your comments?

The child continued to have severe colicky abdominal pain and the bowel sounds disappeared. There had been no recurrence of malena. A Ryle's tube was inserted and antimicrobials changed to ceftriaxone, amikacin and metronidazole. The rash worsened further and by this time had spread on to the gluteal region. The child also started complaining of pain in the ankles. An emergency laparotomy was planned.

**Question III:** What would you have done in such a situation?

Meanwhile the case was reviewed with the attending consultant. He noted that the rash had now involved both the legs and was most prominent on the extensor aspect. He also noted that there was swelling of the right ankle joint and passive movements were painful. He offered a clinical possibility of Henoch Schonlein Purpura.

**Question IV:** What is your opinion?

The surgical team did not agree with this possibility and insisted on proceeding with the laparotomy. The attending pediatrician, however, thought otherwise and suggested conservative management. He advised a course of parenteral steroids and suggested discontinuation of antimicrobials. The parents were in a dilemma as they were being proffered contradictory opinions.

**Question V:** How would you have managed this situation?

The parents opted for conservative management and the child showed gradual improvement. The pain decreased, bowel sounds reappeared and the abdominal distension gradually settled. By day 3 of hospitalization the child had started taking oral feeds. Prednisolone was continued for 10 days. The skin rash, however, persisted and even became confluent. It spread on to the gluteal regions.



## Question VI:

1. Is this the usual clinical course of Henoch Schonlein Purpura?
2. How long would you continue steroids in such a situation?
3. Is use of steroids mandatory in children with HSP having significant GIT involvement? In what situations are steroids indicated?

The child was about to be discharged when the parents noticed that he was passing reddish coloured urine. The urine output had also decreased and the blood pressure had gone up to 140/105 mmHg. Urine examination showed albumin 4+ and a full field of RBCs; blood urea was 250 mg% and serum creatinine was 4 mg%. The attending pediatrician ordered a renal biopsy – it showed mesangial hypercellularity, synechiae formation and cellular crescents in 60% glomeruli. The interstitium was normal. On immunofluorescence there were deposits of IgA in the mesangium with scattered C3 deposits. There was no deposition of IgG and IgM.

A diagnosis of mesangioproliferative glomerulonephritis with crescents was made (ISKDC Grade III). The attending pediatrician and the nephrologist saw the child jointly – there was considerable debate regarding the treatment regimen to be adopted.

## Question VII:

1. How would you have managed this situation?
2. What prognosis would you have given to the parents for the short term and for the long term?

## CASE 3

A 6 year-old boy presented to the Casualty Services with a history of generalised seizures, tonic-clonic in type, associated with loss of consciousness. There had been no past or family history of seizures.

On direct questioning it was found that he had been running low-grade fever for the last 2 months and had been quite listless and lethargic.

He had also been frequently complaining of 'aches and pains' in the limbs.

He had had severe abdominal pain since the day before admission.

On examination he was found to be comatose and had right facial nerve palsy. Fundus examination showed narrowing of the arterioles with silver streaking and venous nipping. Few hemorrhages were also seen. Blood pressure was 180/110 mmHg. There was a peculiar skin rash over the legs which the parents had noticed since morning that day. He passed a black tarry stool in the Emergency Room.

## Question I:

What is your assessment of this child's illness?

He was treated with intravenous diazepam and phenytoin. The seizures did not recur and the child gradually regained consciousness. Blood pressure was controlled with a sodium nitroprusside drip followed by enalapril and propranolol. He continued to have malena. It was decided to subject the child to upper and lower gastrointestinal endoscopy. The attending pediatrician, however, felt that as the most likely diagnosis in this child was polyarteritis nodosa, these procedures were not necessary.



**Question II:** What is your opinion?

It is said that polyarteritis nodosa is one of the most difficult clinical conditions to diagnose in a child.

**Question III:** Do you agree with this statement?  
How do you confirm a diagnosis of polyarteritis nodosa?  
What is the relevance of Ozen's criteria?

A digital subtraction angiography was performed and it showed multiple aneurysms in the renal, celiac and mesenteric arteries.

**Question IV:** 1. What is the diagnostic relevance of these aneurysms?  
2. Are these pathognomonic of polyarteritis nodosa?

It is said that upto 30% patients of polyarteritis nodosa may be Hepatitis B surface antigen positive.

**Question V:** 1. Is this figure applicable for children as well?  
2. Do you think this viral infection can be directly implicated in the pathogenesis of polyarteritis nodosa?

The child was started on pulse methylprednisolone (30 mg/kg/day X 5 days) therapy followed by oral prednisolone (2 mg/kg/day) and cyclophosphamide (2 mg/kg/day). In 3 months time the child had recovered completely. The blood pressure had normalised, fever was passive and there had been no recurrence of seizures. The ESR had normalised and CRP was undetectable.

**Question VI:** What treatment regimen would you choose at this stage and how long would you continue therapy?

It is said that children with polyarteritis nodosa who are Hepatitis B surface antigen positive need to be managed differently.

**Question VII:** What treatment regimen would you recommend for such patients?

## CASE 4

Radhika, a 12 year-old girl was admitted to the Emergency Room with generalised seizures of 5 minutes duration and associated loss of consciousness. There was no past or family history of seizures. Physical examination showed that the radial pulses were barely palpable. Blood pressure in lower limbs was 180/110 mmHg. There were no focal deficits.



- Question I:
1. What are your clinical possibilities?
  2. How would you confirm your diagnosis?

Fundus examination showed ischemic changes.

- Question II: What is your interpretation of this finding?

Aortography showed an irregular contour of the aortic arch and descending aorta and narrowing at the level of subclavian arteries.

- Question III: Is this pathognomonic of Takayasu arteritis?

Further investigations revealed that the tuberculin reaction was strongly positive

- Question IV: How would you interpret this test in the given clinical situation?

The patient was started on anti-hypertensive medication and oral steroids at 2mg/kg/day.

- Questions V:
1. How do you titrate anti-hypertensive medications in this condition?
  2. How long would you continue steroids?
  3. Is there a role for other immunosuppressive agents?

"The 1<sup>st</sup> RCIAP National conference acknowledges contribution of all the above cases 1 to 4 by faculty member, Surjit Singh, from **PGI, Chandigarh** and thanks him for his valuable participation."

## CASE 5

A 2 year-old male child was brought to us from Bardoli village, district Surat in the month of January 2003. His chief complaints were:

- Fever since 40-45 days, high- grade, intermittent.
- Swelling of both feet and hands since 10-12 days.
- Swelling of both knees, more in the right knee since last 5 - 7 days.

The child was unable to walk due to the painful swellings.

A blackish discoloration has appeared in foot, palms and in thigh areas since 5-7 days.

There was no significant past history.

- Questions I:
1. What are the differential diagnoses you would keep in mind?
  2. What more do you want to know on history?



**O/E:** A toxic and irritable child with tachycardia. BP was 130/90. There was non-pitting edema on the feet and palms with gangrenous discoloration of these areas. Right knee joint effusion was evident. There was no organomegaly.

**Questions II:**

1. What more can be looked for on examination?
2. Will your differential diagnosis change after examination?
3. Which relevant tests will confirm your diagnosis?

Investigations: WBC: 25,600- polymorphonuclear predominance, Platelets: 6,80,000

ESR; 38 mm, CRP: 42 mg/dl, LFT: SGPT: 60, bilirubin-normal, Renal functions, XRC, USG (abd) were normal. ANCA, RF, ANA were done.

**Questions III:**

1. What would you expect ANCA, RF factor, ANA to be?
2. What other investigations will complete the work up ?
3. Which is the commonest vasculitis in India?

The child was treated with anti-inflammatory drugs and became asymptomatic in 30 days, was discharged and has been lost to follow-up.

**Questions IV:**

1. How would you manage this child?
2. What would be the follow-up markers for the disease and its management?
3. How would you like to counsel the parents?
4. What is his long-term prognosis?

“The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by Ketan Shah from **Surat** and thanks him for his valuable participation.”

## CASE 6

This patient presented with

- high-grade, continuous fever since 3 weeks
- gangrene of the tip of the left middle finger; subcutaneous lesions on the extremities and face, and arthritis, all since 2 weeks
- abdominal pain since 1 day.

Digital gangrene subsequently progressed to involve 5 other fingers as well.

The fever was initially associated with bilateral submandibular swelling that subsided in 3-4 days. Anorexia, malaise, irritability, aphthous ulcers and pain and tenderness in the legs were present. Past history was not significant except for a history of mild intermittent bronchial asthma since the past 1 year.



Questions I:

1. Any thoughts on the various possibilities ?
2. What was the submandibular swelling? Is it known to occur in this condition?
3. What are the aphthous ulcers due to?

On examination, hypertension was noted, so also edema feet, livedo reticularis on both cheeks, subcutaneous nodules on the lower limbs and atrophic blackish scars on the dorsa of both feet. Circumscribed dry gangrene was present on the distal phalanx of the left middle finger.

Investigations revealed marked leucocytosis, a high ESR and positive CRP, and RBBB on the EKG. Renal parameters, absolute eosinophil count, ANA, dsDNA and abdominal ultrasound were normal. Skin biopsy showed evidence of polyarteritis nodosa.

Questions II:

1. What are the gold standards for diagnosis of PAN?
2. What will be the immediate and long-term management?

“The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by the Pediatric Team (Arpita Gogoi, Radha Ghildiyal, Mrunalini Chavarkar, Shivanand Medar) of **BYL Nair Hospital, Mumbai** and thanks them for their valuable participation”

OVER TO SESSION 4...

IMPORTANT- NOTE CHANGE IN VENUE

The venue has been changed from  
Tata Hospital auditorium to  
**NEHRU SCIENCE CENTRE**



## SESSION : 4

### CASE 1

An 8 year-old boy comes to the outpatient clinic with inability to get up from the bed for the last several weeks.

He was well 6 months ago when the parents first noticed that he was finding it increasingly difficult to get up from the squatting position, to comb his hair and to climb stairs. He also had difficulty dressing himself. These complaints were progressive.

He had also been having episodes of severe abdominal pain, diffuse in nature, lasting for a few hours for the last couple of weeks. These episodes had also been progressively increasing in severity and frequency.

In addition he had been running low-grade fever for the last 4 weeks.

There was considerable malaise and loss of appetite.

The child also complained of occasional swallowing difficulty and regurgitation of fluids through the nose.

There was no relevant past history. His father was a farmer and he had an older female sibling who was alive and well. The family had a pet cat.

The weakness gradually progressed over the subsequent 6 months. He was seen by the family physician, who confirmed the findings of proximal muscle weakness but did not find any other abnormality. He was advised further investigations but the parents were unable to take him to a referral center because of personal reasons.

#### Question I: What are the likely clinical possibilities?

When he reported to our hospital he was found to have profound muscle weakness.

He was:

- virtually bed ridden and had to be carried to the toilet.
- unable to rise from a squatting posture or get out of bed without assistance.
- unable to maintain a sitting posture and could not hold his head upright.

He could still write quite well and could button and unbutton his clothes.

There were contractures at the knees.

There was marked muscle tenderness.

#### Question II: What is your clinical assessment now?

Systemic examination revealed a pulse rate of 84/minute, respiratory rate of 48/minute and a blood pressure of 130/94 mmHg (>95<sup>th</sup> centile). He was afebrile. He had lymphadenopathy in the cervical, axillary and inguinal regions: 1-2 cms, discrete, nontender. Chest examination revealed fine crepts and rhonchi. The cardiovascular system was unremarkable. Central nervous system examination was normal.



- Questions III:
1. How would you interpret these findings?
  2. Why is he tachypnoeic?
  3. How do you explain the hypertension?

Examination of the skin showed several interesting findings.

- Question IV:
1. What would you be specifically looking for?

There was a heliotrope rash.

- Questions V:
1. Is there a differential diagnosis for this rash?
  2. Why does it have a predilection for the eyelids?
  3. Does it disappear on treatment?

There were Gottron's papules.

- Questions VI:
1. Where do you normally look for them?
  2. Can you expect to see these on the feet?
  3. Do they occur in any other disease condition?
  4. Do they disappear on treatment?

There was a prominent photosensitive rash.

- Questions VII:
1. How does one define photosensitivity?
  2. Is photosensitivity a common finding in children with juvenile dermatomyositis?
  3. How do you treat photosensitivity?
  4. Does this disappear on treatment?

There were nailfold telangiectases.

- Questions VIII:
1. How do you look for these?
  2. In which other condition can you get these findings?

Based on these clinical findings a diagnosis was made – no other differential diagnosis was considered.

- Questions IX:
1. What was the diagnosis made?
  2. Would you keep a differential diagnosis?
  3. Why is dermatomyositis more common than polymyositis in children?



The child was investigated.

Hemoglobin	Total WBC count	Differential WBC count	Platelet count	ESR	Peripheral blood film
7 gm%	16000/cu.mm.	P <sub>76</sub> L <sub>20</sub> E <sub>4</sub>	750,000/cu.mm.	100 mm/hr	Normocytic, normochromic

Question X: What is your interpretation of this hemogram?

RFT	LFT	LDH	Cr.Kinase	Aldolase	CRP	Calcium	PO4
BUN and S. Cr were normal	S.Bil: normal SGOT- 480; SGPT: 240	685(↑)	895 (↑)	4 U/L (1.2-8.8)	4000 (67-1800 ng/ml)	10 mg% (S.Na <sup>+</sup> and S.K <sup>+</sup> were normal)	5 mg%

Question XI: What can one conclude from these findings?

Toxoplasma serology	Rheumatoid factor	Antinuclear antibody	Anti-Jo-1	Serum Ig	C3	HLA typing
1:800 EU (N= <800)	Negative IgG normal	Negative	Negative	IgA, IgM,	104 mg%	B8 DR3

Question XII: Do you think that the raised toxoplasma titres are significant?

**Urine examination:** normal.

X-ray chest showed streaky infiltrates and mild cardiomegaly. Electrocardiography revealed evidence of left ventricular hypertrophy and ST depression. Echocardiography showed systolic dysfunction with an ejection fraction of 50% (N=62-84%).

Question XIII: What are your comments?

It was decided that electromyography needed to be carried out.

Questions XIV: 1. Which site would you select?  
2. What would you expect to see?

Electromyography revealed myopathic motor units with denervation potentials, spontaneous fibrillations and high frequency repetitive discharges. The nerve conduction velocity was normal.

Questions XV: 1. How do you interpret these findings?  
2. Can one get a normal electromyogram in a patient with JDM?  
3. Under what situations can one expect to get a predominantly neuropathic rather than a myopathic pattern?



The patient was then subjected to an MRI scan.

- Questions XVI:
1. Do you think this investigation is necessary in such a situation?
  2. Should it be carried out in all children with juvenile dermatomyositis?

The MRI scans revealed fibrosis, atrophy and fatty infiltration in proximal muscles of hip on T1 weighted images and a hyperintense signal on T2 weighted images.

- Question XVII: How does one interpret these findings?

The patient was discussed on the ward rounds. The residents wanted to do a muscle biopsy but the attending consultant disagreed.

- Questions XVIII:
1. What is your opinion?
  2. In which situations would you consider muscle biopsy to be mandatory in this condition?
  3. Which muscle should be selected and what are the typical findings?

After obtaining the laboratory reports, a final diagnosis of Juvenile Dermatomyositis was made.

- Question XIX: What is your opinion on Bohan and Peter criteria?

He was started on therapy: an initial pulse of intravenous methylprednisolone-30 mg/kg/day was given for 3 days followed by oral prednisolone 2 mg/kg/day. The consultant also decided to start oral weekly methotrexate (20 mg/m<sup>2</sup>/week).

- Questions XX:
1. What are your views on this starting regimen?
  2. How would you taper therapy?
  3. What would be the duration of therapy?

The parents of this child were very worried.

- Question XXI: How would you counsel such a family?

On follow-up he presented with complaints of skin nodules on both arms and over the abdomen. There was ulceration over some of these nodules associated with extrusion of small whitish flecks.

- Questions XXII:
1. What is your likely diagnosis?
  2. How would you manage this condition?



At the end of therapy 2 years later, the muscle power had improved significantly. However, the parents noticed that his physical appearance had changed considerably and this was causing them distress. There was marked hirsutism.

- Questions XXIII:
1. What do you think is the likely reason?
  2. How would you manage this condition?

“The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by Surjit Singh from **PGI Chandigarh** and thanks him for his valuable participation.”

## CASE 2

**N**, a sweet young girl of 6 years, has been a rather sick child, with a prolonged period of sickness.

She presented to the department of Pediatrics, KMC Manipal during September 2000 for complaints of fever followed by pain, early morning stiffness, restriction of movements and swelling of multiple joints.

Her past history revealed a similar illness starting at the age of 4 years that initially had lasted for a duration of two to three months. At that time her ANA and anti ds-DNA were positive. Child was not on any treatment and symptoms gradually subsided. **A repeat anti ds- DNA** after three months was negative. This was followed by three similar episodes over the next one year. She also had macular erythematous rashes all over the body during the episodes of fever.

On examination, her height and weight were < 10<sup>th</sup> %tile. Her blood pressure was 130/100 mmHg (> 95<sup>th</sup> percentile). Dark and erythematous macular lesions were present all over her body. Heliotropic rashes were noticed over both eyelids. Bilateral ankle joints and metatarsophalangeal joints were swollen. Mild restriction of movements was noticed bilaterally around knees, wrists and small joints of hands. Pain and weakness was noticed in proximal muscles of limbs. A large firm liver and a small spleen were palpable. Other systems were normal.

- Questions I:
1. What will be your clinical impression?
  2. ANA / ds DNA - comment on sensitivity, specificity and pitfalls?

Investigations:

- normal counts with high ESR (110mm/hr),
- mild elevation of liver enzymes, normal RFT.
- Serum ANA was positive (184u/ml)
- Anti ds DNA was negative.
- RF test was positive (10 IU/ml)
- serum CPK level was elevated (336 IU/dl).
- X ray of knee joints was normal.

- Questions II: How would you manage this child?



She responded to our management with a reduction of ESR to 22mm/hr. Serum ANA repeated was still positive. Child continued to be asymptomatic with adequate growth and normal activities

After 8 months of this treatment, she started developing subcutaneous swellings over multiple sites on the body, which were indurated but not painful. The skin over them could not be pinched. Otherwise she was largely asymptomatic. Skin biopsy identified the condition. Direct immunofluorescence showed features s/o SLE.

**Question III:** What do you think the skin biopsy showed? In which rheumatological conditions is this seen?

Child was asymptomatic, on medications for the next year with persistence of swellings. After one year, she started developing increasing lesions and was started on penicillamine. C<sub>3</sub>, S.ANA, S.Anti ds-DNA, RF tests repeated were negative. After one month, one of the lesions ruptured, forming a non-healing ulcer. ESR was also elevated at this visit (125mm/hr). She has subsequently been started on colchicines but her skin lesions do not seem to be responding and are still increasing

**Questions IV:**

1. Would you like to investigate her further?
2. Outline the role of penicillamine and colchicine here?

"The 1<sup>st</sup> RCIAP National conference acknowledges this case contribution by Shrikiran. A. Hebbar and Vijaya Shenoy from dept. of Pediatrics, **KMC Manipal** and thanks them for their valuable participation."

OVER TO NEXT PART OF SESSION 4...

## Thank you!

**IAP Rheumatology chapter gratefully acknowledges  
the sponsorship and participation of the following corporates:**

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## SESSION : 4

### CASE 1

13 year-old A resides in New Jersey with her family. They are a close-knit family with very caring parents. In 1997, she first complained of ankle pain that was diagnosed as tendonitis and treated with NSAIDs.

A year later she developed bilateral ankle pain that lasted for 7 days. Again tendonitis was diagnosed and she underwent physiotherapy for 8 weeks after which she was better. During both these episodes, there were no signs elicitable.

In 2000, she suffered from severe debilitating left ankle pain that forced her to use crutches and prevented daily activities including schooling. She was thoroughly investigated and all investigations were normal. Notable were a normal MRI and acute phase reactants. Her pain continued to be refractory to physiotherapy, NSAID's. It started improving gradually after 2 months. This was followed by a pain-free period of 1¼ years.

Pain then recurred and this time spread from left knee? left shoulder, progressing over a week with no signs and normal X-Rays. Her ballet, swimming lessons were gradually dropped and she reduced her physical activities. Subsequently she had hip pains, all of which were treated by pain- killers and physiotherapy. She has started having sleep disturbances and is unable to sustain her sleep . She seldom wakes up feeling refreshed.

In school, **A** has been having problems with her gym teacher and now is socially isolating herself. Otherwise, she is a lovely girl who visited us with "a thick file, with a smile."

Her present complaint is an ankle pain and swelling since 5 weeks . The mother states that she has occasionally seen a purplish hue around the ankle joint .At history taking you note that mom is doing most of the talking while A seems almost unconcerned. While talking to you she seems quite comfortable and lifts her leg to the couch effortlessly.

#### Questions:

1. What are your first thoughts?
2. What are you likely to find on physical examination?
3. What investigations are likely to help in arriving at a diagnosis?
4. How will you counsel the parents? What about the longterm prognosis?
5. What are the principles of managing the problems? Will you need help of your colleagues in other fields?

(Case compiled by Rachana Hasija, **Jaslok Hospital, Mumbai.**)

**'This book has a limited print run. No copies will be available at the venue.  
You are thus requested to carry your mailed copy to the conference.'**



<b>FEEDBACK FORM (✓)</b> <b>(LETS DO BETTER!)</b>			
<b>GENERAL</b>	<b>GOOD</b>	<b>FAIR</b>	<b>POOR</b>
VENUE	<input type="text"/>	<input type="text"/>	<input type="text"/>
FOOD	<input type="text"/>	<input type="text"/>	<input type="text"/>
REGISTRATION & GIVE AWAYS	<input type="text"/>	<input type="text"/>	<input type="text"/>
AUDIOVISUALS	<input type="text"/>	<input type="text"/>	<input type="text"/>
CORRESPONDENCE (if applicable)	Prompt <input type="text"/>	Delayed <input type="text"/>	<input type="text"/>
HOUSING NEEDS (if applicable)	Cost effective <input type="text"/>	Could be better <input type="text"/>	Poor <input type="text"/>
<b>SCIENTIFIC</b>	<b>GOOD</b>	<b>FAIR</b>	<b>POOR</b>
CONTENT	<input type="text"/>	<input type="text"/>	<input type="text"/>
FORMAT	<input type="text"/>	<input type="text"/>	<input type="text"/>
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CONFERENCE BOOK	<input type="text"/>	<input type="text"/>	<input type="text"/>

What did you like most about this conference? \_\_\_\_\_  
 \_\_\_\_\_

What did you dislike most about this conference? \_\_\_\_\_  
 \_\_\_\_\_

WHICH POSTER DESERVES THE PRIZE (ONLY ONE) \_\_\_\_\_  
 \_\_\_\_\_